Sarcomatoid Urothelial Carcinoma of the Renal Pelvis with Extremely Aggressive Clinical Behavior

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Sarcomatoid urothelial carcinoma is a rare malignancy with a poor prognosis. We experienced a case of sarcomatoid urothelial carcinoma of the renal pelvis with extremely aggressive clinical behavior. An 81-year-old woman underwent a laparoscopic radical nephroureterectomy to remove a 4.5x3.1 cm sized localized left renal pelvis mass. The mass was pathologically confirmed as a sarcomatoid urothelial carcinoma. Although the operation was successful, the patient died 2 months postoperatively with widely metastatic disease. (Korean J Urol 2009;50:812-815)

Key Words: Kidney pelvis, Carcinoma

Sarcomatoid urothelial carcinoma of the renal pelvis is an uncommon variant of urothelial carcinoma that has both malignant epithelial and mesenchymal components. This is a rare malignant tumor. Only 1 case of primary sarcomatoid urothelial carcinoma of the renal pelvis has been reported in Korea. This tumor is very difficult to diagnose, and immunohistochemistry is helpful for a correct diagnosis. The clinical behavior of this tumor is aggressive. We experienced a case of a sarcomatoid urothelial carcinoma of the renal pelvis with extremely aggressive clinical behavior in an 81-year-old woman.

CASE REPORT

An 81-year-old woman presented with a 2 week history of painless gross hematuria. She was healthy with no medical problems and denied smoking and drug abuse. No mass was palpable in the abdomen during a physical examination. Urine cytology was suspicious of transitional cell carcinoma. Abdominal ultrasonography demonstrated dilation of the pelvocalyceal system of the left kidney. In the cystoscopic examination, jetting of hematuria from the left ureteral orifice was observed. Computed tomography (CT) revealed a 4.5x3.1 cm well-enhancing mass in the left renal pelvis and no distant metastasis (Fig. 1). Metastatic evaluation including chest CT and nuclear medicine bone scan demonstrated no sign of disease spread.

The patient underwent a left laparoscopic radical nephroureterectomy with no intraoperative problems. The entrapped specimen was divided into 3 pieces and removed through a 3 cm sized initial port site. Grossly, there was a 4.5x3x2 cm sized exophytic, multilobulated and polypoid mass at the renal pelvis. Histologically, the tumor consisted of neoplastic spindle cells
that partly formed a fascicular arrangement with frequent vascular invasion (Fig. 2). Meticulous sections of the tumor revealed a few foci of urothelial components and merging of the epithelial and spindle cell components (Fig. 2). There was no heterologous element in the meticulous sections from the tumor mass. Immunohistochemical analysis was performed with antibodies against cytokeratin, smooth muscle-specific actin, desmin, and vimentin. The spindle cell elements showed coexpression of both cytokeratin and vimentin and were negative for muscle-specific actin and desmin (Fig. 3). These findings led to the diagnosis of a sarcomatoid urothelial carcinoma without heterologous elements according to the 2004 WHO classification of tumors of the urinary system and male genital organs. The postoperative period was unremarkable, and the patient was discharged home 5 days after the operation. One month after surgery, however, the patient was readmitted because of nausea, poor oral intake, and general weakness. Chest radiography showed multiple small nodules suspicious of

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**Fig. 2.** Microscopic findings of the specimen. (A) Neoplastic spindle cells arising from a high-grade urothelial carcinoma (H&E, x400). (B) Sarcomatoid spindle cells in a fascicular arrangement (H&E, x200).

**Fig. 3.** Immunohistochemical findings of the specimen. (A) Cytokeratin stain shows positive reactions in the sarcomatoid spindle cells (cytokeratin stain, x400). (B) Vimentin stain shows strong expression of vimentin in the sarcomatoid spindle cells (vimentin stain, x200).
DISCUSSION

Sarcomatoid urothelial carcinoma of the renal pelvis is a very unusual malignant tumor, consisting of an admixture of carcinomatous and sarcomatoid components. Histologically, the epithelial component is urothelial, and in some cases the epithelial component cannot be recognized. In the current case, only a few areas of the multiple sections from the tumor included high-grade papillary urothelial carcinoma. Immunohistochemical studies are mandatory for the diagnosis to prove biphasic differentiation such as cytokeratin showing epithelial differentiation and vimentin showing mesenchymal differentiation. Coexpression of vimentin and cytokeratin was demonstrable in the present case. According to the 2004 WHO classification of tumors of the urinary system and male genital organs, heterologous elements including chondrosarcoma, osteosarcoma, or rhabdomyosarcoma express markers appropriate to the type of differentiation; these include S-100, muscle-specific actin, desmin, and myoglobin. The immuno-

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